A Case of Ectrodactyly in a Neonate

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ABSTRACT

Ectrodactyly also known as Split hand/foot malformation is a rare limb malformation with autosomal dominant in heritance with variable penetrance, commonly known as "lobster claw hand". Usually it involves midline clefts of the hands and feet with syndactyly. We report a neonate with ectrodactyly and brief review of literature of condition.

Key words:
Ectrodactyly, split hand/foot malformation, syndactyly

INTRODUCTION

Ectrodactyly or split hand-foot malformation (SHFM) is a rare malformation involving the central rays of the hands and feet. Usually, it affects one to all four limbs randomly. It is associated with syndactyly with oligodactyly and rarely polydactyly leading to "lobster claw" appearance. Its prevalence has been reported to be about 1-9/100000.[1] The deformity is present since birth and maybe associated with other dysmorphic features and anomalies in some syndromes. Non-syndromic patients usually follow autosomal dominant inheritance with variablepenetrance. Syndromic patients have autosomal recessive or x-linked recessive inheritance. Sensorineural hearing loss is common in patient with ectrodactyly with syndromic association. It has also been observed with Aniridia and Alport syndrome.[1] Five different genetic mutations are known to be associated with SHFM. Type I, the most frequent variety, is due to a mutation on chromosome 7.[1] We report a neonate showing ectrodactyly [Figure 1] and syndactyly and polydactyly [Figure 2] of right lower limb.

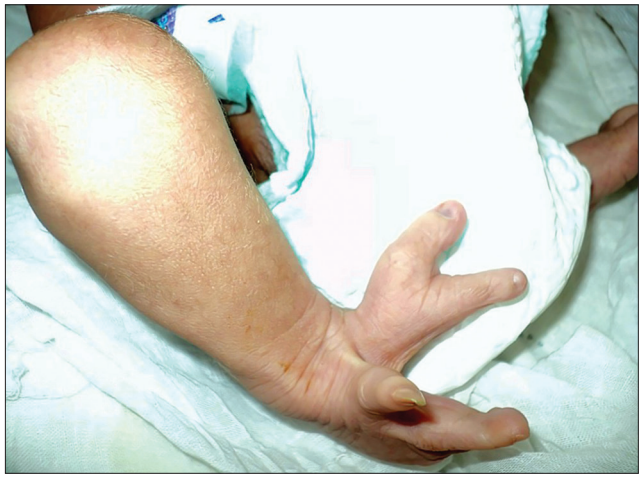

Figure 1: Ectrodactyly of right lower limb

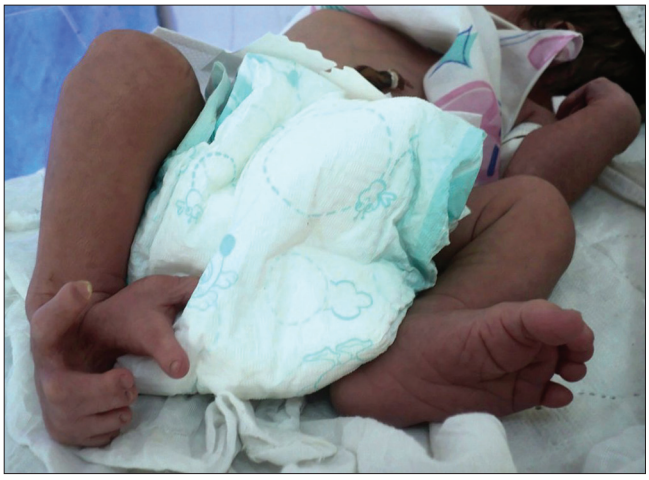

Figure 2: Syndactyly and polydactyly of right lower limb

Ectrodactyly can be treated surgically in order to improve function and appearance. Prosthetics may also be used.[2] Parents should be counseled regarding the possibility of recurrence of the disease in the future siblings and antenatal diagnosis by ultrasonography should be offered.[3]

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